CEREBELLAR TITUBATION; SUNSTROKE SEQUELÆ; SYRINGOMYELIA; TRAUMATIC HYSTERIA.

CLINICAL LECTURE DELIVERED AT THE PHILADELPHIA HOSPITAL.

BY F. X. DERCUM, M.D.,

Clinical Professor of Diseases of the Nervous System, Jefferson Medical College; Neurologist to the Philadelphia Hospital.

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GENTLEMEN,—The first patient I bring before you this morning is a very interesting case. Unfortunately, the data on which to base a diagnosis are wanting. Even as regards the past history we are left badly in the lurch. The man is a German, speaks with considerable difficulty, and presents, unfortunately, very evident mental failure. However, from this unsatisfactory source, we have gathered that in June, 1888, he was in the open air and exposed to the sun. He says that he had a sunstroke. Whether he had been drinking or not we cannot find out. At any rate, it appears that he fell to the ground, was removed to his home, and remained unconscious for several weeks. I have carefully inquired at the address that he gave me, and in the neighborhood in which he tells us he lived, but absolutely without result. I have not been able to discover any of his friends.

As you see, his movements are very irregular. They are, however, not ataxic, but decidedly jerky. Observe, now, the unsteady manner in which he takes hold of this book that I hand him. It is not that the movement lacks certainty so much as it lacks evenness and regularity. I now get him to stand up, and you at once see that his attitude is peculiar (see Fig. 1). He stands with the feet well separated, the arms extended laterally, and the head thrown slightly back. The whole posture is suggestive of the effort of balancing,—of the effort of maintaining his equilibrium. If now I get him to walk, you notice at once that the gait is strikingly abnormal, and yet, if you were asked to classify it, you would meet with some difficulty. Certainly it is not the gait of weakness, nor is it an ataxic or a spastic gait. Note that

FIG. 1.—Probable Cerebellar Titubation. Showing position assumed by a patient.

he still keeps his feet widely separated, and that he moves the feet forward by little, short, jerky steps. Every now and then he stops, sways to and fro, balances himself anew, and then starts afresh. He is apt to fall if not watched, and some time ago suffered from a fractured fibula in consequence. He frequently saves himself by grasping surrounding objects. His gait is eminently a staggering gait, the tendency to fall forward or backward being more marked than the tendency to fall sidewise. It looks for all the world like cerebellar titubation. I can make out no anæsthesia, and the knee-jerks, while diminished, are not absent. There is no nystagmus. His pupils react well. An examination of his eye-grounds reveals that they are hazy and deficient in color. There is no change in the central vessels. These conditions are those of partial optic atrophy, and have become more marked with time.

It would be useless to speculate in detail upon the possible lesion in this man's case. Suffice it for the present to say that he presents cerebellar symptoms, and it is for this reason that, in spite of our scanty history and absence of diagnosis, I bring him before you. I do not think that any of you, once having seen this case, will ever confuse such a gait with ataxia.[1]

It is not impossible that these symptoms are really the outcome of a sunstroke. Sunstroke sequelæ are occasionally not only very persistent, but often very unexpected. This I will illustrate by the next patient. Fortunately for us, he is very intelligent, and gives us the following history:

He is thirty-one years of age, a native of Ireland, and a laborer. Family history good. He himself had never suffered from any illness previous to the present trouble. Had been a moderate drinker, but had never drunk to excess. He denies any venereal taint. Physically, he presents a large frame and looks as though he had at one time been possessed of a fine muscular development.

In July, 1887, while working in the sun, he had a heatstroke. He fell unconscious to the ground and appears to have been very ill. He remembers nothing clearly for some five weeks afterwards. He was told that during the greater portion of this time he had been delirious. While convalescing he had great difficulty in talking; says that at first he could not talk at all; says, also, that he could not move his hands or feet, and that his legs and arms were numb. Gradually, however,

[1] Since the above lecture was delivered the patient died of an intercurrent diarrhœa. The autopsy revealed, in addition to other changes of minor importance, excessive atrophy and softening of the cerebellum.

he began to talk better, and little by little regained power over his arms and legs. In the course of four or five months he could walk a little with a cane. He suffered, however, at times with dizziness. Headache, present at first, had now disapeared. His condition was essentially chronic, and in February, 1888, he was admitted to my wards. Here it was noted that he was exceedingly weak in both arms and legs. He could take a few steps with the aid of a cane, if supported, while his grip on both sides was much below normal and tremulous. Sensation had much improved, so as to be practically normal in the arms and but slightly below normal in the legs. Dizziness was still present at times. Speaking seemed to fatigue him, and it was noticed that the labials and especially the dentals were pronounced very indistinctly. He could not whistle. The cheeks seemed smoothed out and flattened, and there was slight drooping of the lower lip on the left side.

It was further noted that there was slight flattening of the deltoid and supra- and infra-spinatus muscles. Marked fibrillary tremor was present in all of these muscles, as well as in the biceps and triceps. Distinct wasting was, however, not present in the muscles of the arms, forearms, or hands. In the forearms, indeed, fibrillary tremors were not visible; however, if the arms were grasped by the hands of the physician a little below the elbow, a coarse thrill was felt. This thrill became more marked when the patient threw the muscles into contraction by firmly closing the hand. It reminded one very much of the sensation received when resting the hand on the head of a purring cat. On applying the ear it seemed in no way to resemble the ordinary muscle-note, but seemed louder, coarser, and interrupted. Fibrillary tremors were also present in the gluteal region, the muscles of the thigh, and the calf muscles. There was also slight tremor of the lips and marked tremor of the tongue. All the muscles of the trunk and legs appeared small, but wasting, if present, was not excessive. Pain was absent, except in the lower dorsal and lumbar region of the back, where dull and persistent aching was complained of. The knee-jerks were not changed, unless they were slightly below normal. Electrically, there was a slightly-lessened response, especially in the muscles about the shoulders. The sphincters, it should be stated, had never been involved. The hands and feet were cold and livid.

He was treated by tonics and rest, but little or no change resulted. The pain in the back persisting, the actual cautery was freely applied to the spine. The effect upon the pain was almost immediate. A month later, some traces of pain still persisting, the cautery was applied

a second time. From this time on a decided improvement ensued; the pain disappeared entirely, and there was a steady increase in the muscular strength. By the following June (1888), this increase was very decided, while all traces of anæsthesia had disappeared, and the tremor of the hands had become very slight.

As the man stands before you, the peculiarity of his face is very striking. The countenance does not play with emotion as he talks, but it certainly is not fixed, as, for instance, we have seen it in paralysis agitans. Again, it is undoubtedly smooth, but it is evidently not the smoothness which is seen in dementia. The characteristic of the face is undoubtedly the flattening, which, in turn, is the direct result of wasting. Looking closely, we find that the lips are still slightly tremulous, while in the back and shoulders fibrillary tremors are still seen here and there. Again, if I grasp the forearm I still recognize the "purring" sensation.

This case illustrates a rare and exceedingly interesting sequel of sunstroke. We all are familiar with the fact that sunstroke may give rise to inflammation of the membranes of the brain, as well as to destructive changes in the cortex itself. This man, we should remember, was for weeks delirious and unconscious, and afterwards suffered for a time from headache and vertigo. At the same time that these brain-symptoms existed, signs of spinal-cord involvement were also present. It would indeed seem as though the disturbing influences following the heatstroke had spent their force rather on the cord than on the brain or its membranes. There can, in fact, be little doubt that this man suffered from a more or less marked poliomyelitis. The case, we may say, can be explained on no other ground.

Here is another patient also illustrating a sunstroke sequela involving the cord. Not to consume too much time, I will merely mention the principal points of his history. He is some forty-odd years of age, and was in good health up to three years ago. He was at that time in Atlantic City, and in the month of August suffered a sunstroke. After the immediate symptoms passed away he suffered from headache, and two weeks later he noticed weakness of the left leg and in a few days a similar weakness of the right leg; at the same time there was distinct numbness. This condition persisted until, as you see now, the man became completely paraplegic.

Examining the legs more closely, we find that the loss of power is absolute. The knee-jerks are much exaggerated, and ankle-clonus is present and decided. Sensation is diminished but not abolished. The sphincters are intact. Girdle pains, though present, are not

marked. Evidently this man suffers from a myelitis which, though diffused, is most marked in the motor tracts and involves the lumbar portion of the cord.

Leaving now this interesting series of sunstroke cases, let me call your attention to a fourth patient, who presents a very unusual condition. His history is as follows:

C. H., aged forty years, a native of Sweden, unmarried, a laborer. His father died of phthisis; his mother of old age. Has two brothers, both living and in good health. He himself had typhoid fever when twenty years of age. Has had no other illness. Denies venereal history, and has always been temperate in his habits.

About twelve years ago, while working in a saw-mill, he did some heavy lifting and strained his back. He was obliged to go to bed for a day, but afterwards felt no evil effects. Three years later he began to suffer from pains in the legs and band-like pains about the lower part of the chest. These pains gradually grew worse, and soon his walking was interfered with. He was after a while obliged to use crutches.

He was admitted to the hospital in January, 1883. He was unable to walk, but could sit up in a chair. Since then his condition has gradually changed for the worse; but considering the length of time that has elapsed, the change has been comparatively slight.

Without pausing to read to you the further notes of his case, let me call your attention to his present condition. You notice, as he lies before you, that there is some wasting in the legs; further, that the right leg is drawn up towards the trunk. When I ask him to move his feet or his legs, you see that he is unable to comply; and when I take hold of the right leg and attempt to extend it, we find that the muscles are firmly contracted. The leg is partly flexed on the thigh, and the thigh on the abdomen. Looking at the left leg, we find that it also is stiff, but not contracted as its fellow. Testing the knee-jerks, we find on the left side that it is markedly exaggerated; on the right side, however, owing to the fixed position of the limb, no movement can be elicited. Ankle-clonus is also present on the left. The skin reflexes also are very much increased.

Examining now the cutaneous sensibility, we find that it is absolutely lost in both legs, and, further, that this loss extends upon the trunk. It is not, indeed, until I reach with the æsthesiometer the level of the nipple on the right side and the level of the umbilicus on the left, that he begins to respond. You notice further that his answers lack certainty until I fairly pass these levels. His answers are at first

FIG. 2.—Syringomyelia. Extensive arthropathy of the right shoulder.

slow and hesitating, probably because the sensory impulse is feeble and delayed. Testing now his sense of temperature in the parts of his body in which cutaneous sensibility still appears to be present, we find that he fails absolutely to recognize the difference between hot and cold objects over the right arm, right shoulder, right side of neck and face. On the corresponding parts on the left side the temperature sense is well preserved. Some analgesia is also present over the right arm. Constricting pains are still present in the chest. Both sphincters are paralyzed. There is incontinence both of the bladder and rectum. Lastly, turning him upon the side, we find the scar of an old bed-sore on the buttocks.

There is still another and a most remarkable condition present, and on account of its rarity I bring this patient before you.

As the patient holds up his arms you notice at once (see Fig. 2) that the right shoulder presents a very peculiar appearance and is in marked contrast with the left, which is normal. When I ask you to express an opinion, one of your number replies that the arm is dislocated. This is perfectly true, but it is not the whole truth. If I take hold of the humerus and roll it in its socket, I feel that the joint is very much roughened. It is freely movable in all directions, but the grating is so loud that you can almost hear it where you sit. Our patient, in the mean while, does not give the least evidence of pain, and even when the movements I perform are anything but gentle, it fails to hurt him. You have here, then, an instance of extensive disease of a joint which is absolutely painless. If I examine it more closely, I find that the capsule contains fluid. I fail, however, to make out any loose fragments of bone.

The disease of this joint is trophic. It is evidently identical with the affection not infrequently found in locomotor ataxia, the description of which we owe to Charcot, and of which I showed you several instances last year. Like other structures, the bones depend for their growth and nutrition upon the integrity of certain nervous centres. In locomotor axtaxia in which we have sclerosis of the posterior root-zones, disease of the joints is exceedingly liable to occur. That, however, it does not occur in every case proves that there is yet some unknown factor which, in addition to the disease of the posterior root zones, determines the affection. Curiously enough, there is no change in the joints of the lower limbs.

One of the curious facts of this disease of the joints is the occasional suddenness of its onset. Two years ago, in this man's case, he noticed a sudden swelling of the shoulder. It was painless, but the

affection persisted from that day on. When these cases are examined post mortem, it is found that the cartilage of the bones has been eroded, that here and there bone has been absorbed, but especially that there has been irregular and often extensive osseous deposit. Occasionally the capsule of the joint ruptures and permits the escape of the contained fluid into the surrounding tissues. This actually happened at one time in the present case, and the fluid permeated the inter-muscular septa, giving rise to quite a troublesome cellulitis. This has now, however, disappeared.

The case before us is probably one of syringomyelia; that is, we have here an instance in which the spinal cord has probably undergone extensive destruction by reason of great proliferation of the neuroglia and the subsequent formation of a cavity or cavities. The fact that we have here associated muscular wasting with anæsthesia and contracture, together with loss in certain areas of the temperature sense, points strongly to the presence of this disease. Further, the occurrence of a Charcot joint is another fact in harmony with this view. Although rare in syringomyelia, it has been observed before. Again, to suppose our patient suffering from some other disease of the cord, say a myelitis, would involve many difficulties, and especially would leave the remarkable condition of the shoulder-joint unexplained.

Without pausing to devote further time to this interesting case, let me call your attention to the following, which is also very remarkable. The history is briefly as follows:

D. J. F., male, aged thirty-three, married, a druggist by profession, tells us that both his parents are living and well; that his maternal grandfather became insane at the age of eighty-one and died at eighty-three; that he has two brothers, one two years older than himself and perfectly healthy, and another two years younger and "chicken-breasted." No sisters.

He himself had been perfectly healthy until some twelve years ago, when, according to his story, he had a narrow escape from death by lynching, in the Northwest. He had been seized, accused of murder, and forthwith hanged to a neighboring tree. In their effort to make him confess, the lynching party had raised and lowered him a number of times, and finally left him suspended. In the nick of time the mistake was discovered and he was cut down. Long before this unconsciousness had supervened. He was taken to the hotel and put to bed, where he remained under medical attendance for thirteen weeks. He does not remember how long he was unconscious. All he remembers is suffering from a confused sense of horrible and overwhelming

fear which persisted for many weeks, during which the frightful drama was being enacted over and over again. When he improved he suffered a great deal from nervousness: could not get to sleep at night; would have frightful dreams; would wake up with his heart beating very rapidly; was very weak.

After he began to be around, even the slightest noise or a person speaking to him suddenly would make him start and tremble. Little by little he got better, but was not himself at any time. After a while he tried to work. He drifted from one thing to another. He at one time set type, at another acted as reporter for a Western paper, and finally, four years later, found himself in Denver, where he married. This he now bitterly regrets, and thinks, on account of his physical condition at the time, to have been very injurious to him. His general nervous condition persisted, and after a time he began to get weak in the legs. He had been married four years when this weakness became sufficiently well marked to interfere with his walking. He began to stump his toes and would occasionally fall. This difficulty steadily increased for a year, when he came to Philadelphia for treatment. He has been in various hospitals,—the Polyclinic, the Infirmary for Nervous Diseases, the University Hospital, and finally in our own wards. His symptoms steadily persisted until they attained their present severity. He was variously regarded by his physicians as a malingerer, as hysterical, as a case of organic disease. Whatever interpretation may be placed upon him now, of one thing I feel certain, namely, that he is not a malingerer. He all along had hysterical symptoms, and I remember well that at the University Hospital, where I also had the opportunity of seeing him, it was demonstrated beyond all cavil that he possessed more power in his legs than he admitted, or perhaps himself believed, and also that a very curious reflex was developed in his case upon suggestion. However, there remained always a substratum of genuine symptoms that not only failed to disappear, but actually became more and more pronounced.

Let us look at the man as he sits in the chair. I will throw back his head, and at once you behold the horrible scar made by the rope,—a cowboy's lariat. This wound he tells us was fully eighteen months in getting well.

I ask him to rise, and you see that he is almost completely paraplegic. His knee-jerks are much increased, and there is some, though not very marked, rigidity of the limbs. Sensation is slightly obtunded, though not lost. He has no constricting pains, no paralysis of the sphincters. His grip is weak, but the arms are otherwise unaffected.

His pupils are large and very mobile. His features are good, but rather emotional in type.

Practically it matters very little whether this man has actual organic disease of the cord or whether his affection be simply hysterical. It was for a long time considered hysterical, and every method of treatment, every resource, was exhausted without avail. I myself incline to the view that his case is in part organic, though I admit that this cannot be absolutely demonstrated. Certainly his case resembles a lateral sclerosis. That he is also hysterical there can, I think, be no doubt. His hysteria is traumatic, and exceedingly interesting.

You are all familiar with the fact that hysteria often follows fright, especially if coupled with actual mechanical injury, such as we find in railway accidents. Certainly in the present case there is abundant cause furnished by the man's history to explain his condition.